

An uncommon cause of miliary pattern of pulmonary nodules—diffuse pulmonary meningotheliomatosis

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Abstract

Pulmonary meningothelial-like nodules are benign lesions that are often incidentally detected in surgically resected lung tissue. These nodules are usually asymptomatic and single. Rarely, they present as diffuse micronodules similar to the miliary pattern seen in tuberculosis or metastatic cancer. While diffuse meningothelial-like micronodules are usually benign, it is important to include this condition in the differential diagnosis of patients presenting with diffuse micronodules. We present the case of a 74-year-old asymptomatic female referred to the pulmonary clinic for evaluation of incidentally detected diffuse bilateral pulmonary nodules. A transbronchial biopsy established a diagnosis of diffuse pulmonary meningotheliomatosis, obviating the need for further invasive workup. She remains stable after more than 2 years of follow-up.

Introduction

Minute pulmonary meningothelial-like nodules (MPMNs) are common incidental findings in lung biopsy specimens. MPMNs are usually asymptomatic, and may be solitary or multiple. Rarely, these lesions are so numerous that they resemble the miliary pattern on imaging and can be mistaken for metastatic malignancy or infectious diseases such as tuberculosis. Here we describe a case of diffuse pulmonary meningotheliomatosis (DPM) presenting with a miliary pattern.

Case Report

A 74-year-old, non-smoking female was referred to the pulmonology clinic for evaluation of pulmonary nodules incidentally detected on an abdominal computed tomography (CT) scan. Her past medical history was notable for hypertension, dyslipidaemia, prediabetes, and well-controlled asthma. She denied dyspnoea or cough. Review of systems was positive for significant anxiety and weight loss of 6 pounds since being informed about the CT findings. Laboratory results including a complete blood count, kidney function, and electrolytes were within normal limits. A chest CT showed innumerable small solid and ground glass nodular opacities, 5 mm or less in diameter,

throughout the lungs in a predominantly centrilobular distribution (Fig. 1). Some of the nodules demonstrated central cavitation. A repeat CT chest imaging performed 8 weeks later was unchanged. She underwent bronchoscopy. Bronchoalveolar lavage and transbronchial lung biopsy samples were obtained. Bronchoscopy revealed no airway abnormalities. The bronchoalveolar lavage fluid showed no tumour cells or microorganisms. The transbronchial biopsy demonstrated collections of cytologically bland epithelioid cells with occasional cytoplasmic intranuclear pseudoinclusions within the interstitium arranged in a distinctly nested, whorled pattern (Fig. 2). Immunohistochemical stains were strongly positive for epithelial membrane antigen and vimentin, and negative for pancytokeratins, Thyroid Transcription Factor 1 (TTF-1), synaptophysin, chromogranin, S100 protein, and smooth muscle actin. These results supported the diagnosis of MPMNs. Pulmonary function tests revealed normal spirometry, lung volumes, and diffusion capacity. She continued to be asymptomatic on follow-up. Repeat CT imaging, more than 2 years after her initial chest CT, showed stable bilateral lung nodules.

Discussion

It has been more than half a century since MPMNs were first described by Korn et al. in 1960 [1]. However, data is

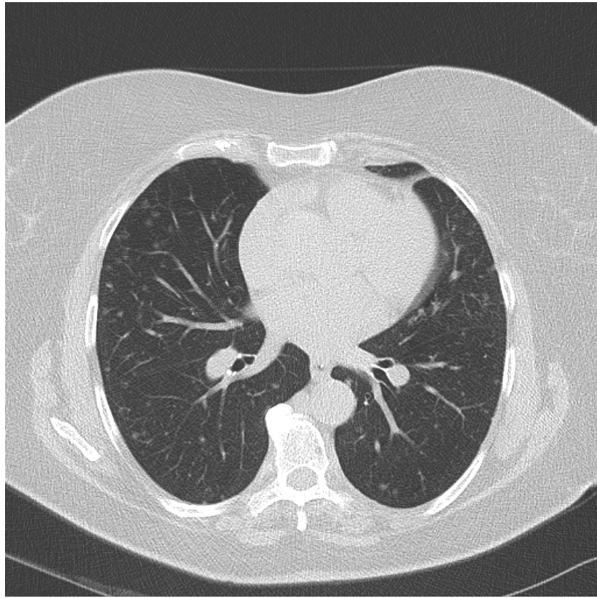


Figure 1. Bilateral diffuse subcentimetre pulmonary nodules caused by diffuse pulmonary meningotheliomatosis.

still emerging regarding their aetiology and pathogenesis. Studies evaluating lung biopsy or lobectomy specimens and autopsies have reported prevalence between 1.1 and 13.8% and 0.07 and 4.9%, respectively [2]. The higher prevalence in non-autopsy series may be due to the difference in methodology, the number of slides examined, or the higher prevalence of coexisting lung disease. This observation suggests that underlying lung disease may provide a stimulus for MPMN growth. Some MPMNs show

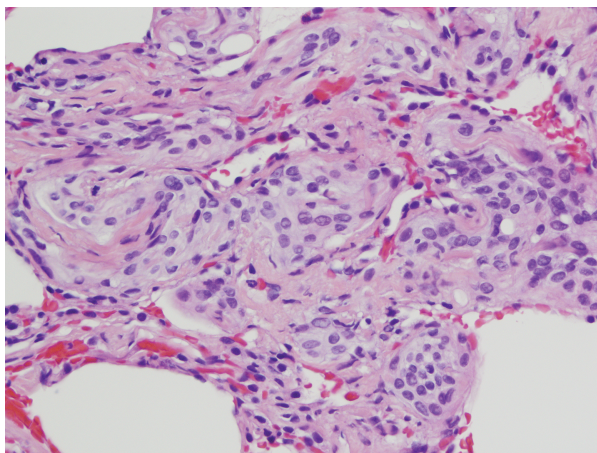


Figure 2. Meningothelial-like nodules in transbronchial lung biopsy specimen. Photomicrograph representing the small, well-circumscribed, whorled-appearing nodules comprised of bland oval cells. The nodules are located within the alveolar septa, unrelated to the pleura or the interlobular septum and are surrounded by small blood vessels.

monoclonal expansion, whereas others are polyclonal, leading investigators to conclude that MPMNs are reactive rather than neoplastic [3].

MPMNs are usually asymptomatic and are generally detected incidentally. MPMNs may be single or multiple. Histologically, these nodules consist of nests of moderately sized elongated cells with oval nuclei, finely granular chromatin, inconspicuous nucleoli, and abundant granular and eosinophilic cytoplasm with indistinct cell borders. Immunohistochemical studies show that cells comprising MPMNs stain positive for epithelial membrane antigen, vimentin, and progesterone receptor [4]. Niho et al. reported that half of MPMNs showed immunoreactivity for the progesterone receptor suggesting a role of sex-steroid hormones in the pathogenesis [3]. MPMNs are most commonly seen in sixth and seventh decades with a notable female preponderance [1,2,4].

MPMNs may be distributed diffusely in both lungs. This condition has been termed “diffuse pulmonary meningotheliomatosis.” Radiologically, DPM usually manifests as diffuse, multiple nodules in both lungs with the size ranging between 100 μ m and 11 mm. Cavitation in DPM nodules is a rare finding and can erroneously suggest granulomatous or infectious processes.

The clinical significance of DPM is unclear. Knowledge about DPM is limited to case reports. Patients are usually asymptomatic as in our case. Fatigue and dyspnoea have been reported, although comorbidities such as ischaemic heart disease, pneumothorax, and lung cancer may explain the presence of symptoms [3]. DPM is not consistently associated with a prior smoking history [3,5].

The radiographic presentation of DPM can mimic several conditions with grave prognoses including metastatic malignancy and miliary tuberculosis. Tissue sampling is important to establish a diagnosis. Most cases are diagnosed with a video-assisted thoracoscopic surgery (VATS) or transthoracic lung needle biopsy. Diagnostic tissue was obtained with transbronchial biopsy in our patient. One additional case report used a similar approach [5]. This highlights the potential value of a less invasive approach to establishing a DPM diagnosis. Management of DPM is usually conservative with observation and follow-up imaging. In our patient, CT scans repeated at 2, 17, and 25 months showed that the lung nodules remained stable.

Disclosure Statement

No conflict of interest declared.

Appropriate written informed consent was obtained for publication of this case report and accompanying images.

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